Operative Techniques in Thoracic and Cardiovascular Surgery

Valve-Sparing Konno and Hypertrophic Obstructive Cardiomyopathy in Children

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Introduction

Septal myectomy is considered to be the most appropriate surgical treatment for patients with hypertrophic obstructive cardiomyopathy (HOCM) and severe symptoms that are unresponsive to medical therapy.[1] Extended transaortic septal myectomy is the procedure of choice in most centers. It provides safe and effective relief of left ventricular outflow tract (LVOT) obstruction. A satisfactory procedure includes subaortic myectomy extended to the midventricular level and correction of anomalies of the mitral papillary muscles. Failure to achieve adequate septal resection or to correct associated mitral anomalies may lead to residual obstruction and the need for reoperation.[2] Adequate septectomy must be carried out apically far enough to prevent contact of the anterior mitral leaflet with the septum at the midventricular level. It is also essential to address adequately associated mitral valve anomalies, such as anomalous papillary muscle insertion directly onto the anterior mitral leaflet, extensive fusion of papillary muscles with the ventricular septum, and abnormal insertions of accessory papillary muscles or chordae tendineae onto the ventricular septum.

In the pediatric population with HOCM, LVOT obstruction may develop either in early infancy or, otherwise, later on during adolescence.[3] In both groups, severe LVOT obstruction may increase rapidly and a surgical approach is recommended because high LVOT gradient is recognized as a risk factor for sudden death. Extended transaortic septal myectomy provides satisfactory results, particularly in experienced centers.[1,4] However, the procedure is associated with an increased risk of residual or recurrent obstruction and of iatrogenic injury to the aortic and the mitral valves.[4] In young children, the operation may be technically challenging because of the difficulty of exposure of the intraventricular lesions through a small aortic orifice. Limited visibility may yield to inadequate resection at the midventricular level or inability to address mitral valve anomalies.

The modified valve-sparing Konno procedure, first described by Cooley and Garrett in 1986, is usually used to relieve tunnel or complex subaortic stenosis while preserving the aortic valve.[5,6] The modified Konno procedure provides excellent exposure of the LVOT, even in small children, thus allowing extensive muscular resection as well as correction of mitral valve anomalies. It is thus particularly indicated in severe forms of HOCM.[7] Since 1990, the modified Konno operation is our procedure of choice in children with HOCM.[8,9] Associated mitral valvuloplasty to correct severe systolic anterior motion of the mitral valve is performed in selected cases, using the retention plasty technique, as described by Delmo Walter et al.[10] Implantation of an epicardial cardiac defibrillator is also carried out, if indicated (history of syncope or sudden death presumably related to arrhythmia, very severe ventricular septal thickness, and a positive family history of sudden death) (Figs. 1-11).

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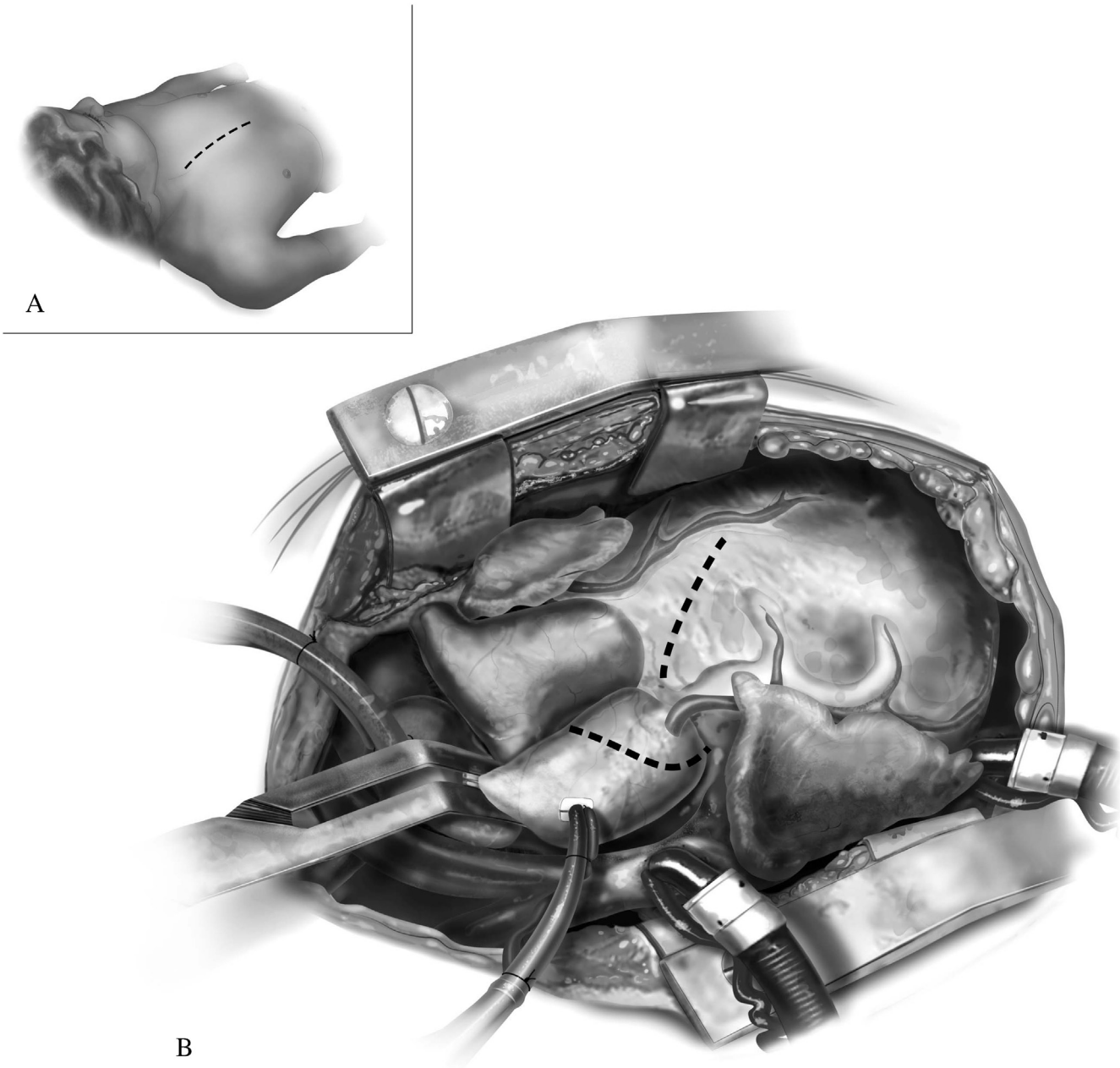

Figure 1 A median sternotomy is performed. Cardiopulmonary bypass is instituted using 2 caval cannulas and an ascending aorta cannula. The aortic cannula is inserted as distally as possible into the ascending aorta. A left vent is placed directly into the left atrium. Cardiopulmonary bypass is carried out in normothermia and using conventional hemofiltration. Because of the presence of severe myocardial hypertrophy, great care should be taken to achieve optimal intraoperative myocardial preservation. Multidose warm blood cardioplegia is our first choice. The initial infusion is given into the ascending aorta; reinfusion doses are administered every 10-12 minutes directly into the coronary ostia. Alternatively, cold histidine-tryptophan-ketoglutarate cardioplegia (Custodiol, Custodiol HTK, Köhler Chemie GmbH, Bensheim, Germany) can be used, particularly in small children, to avoid repeated cannulation of the coronary ostia.

The ascending aorta and the pulmonary trunk are dissected apart as near as possible to the aortic annulus. A diagonal incision is made in the aorta, extended under direct vision, deep into the noncoronary sinus, such as to gain perfect exposure of the aortic valve and the subaortic area. The right ventricle is opened transversely below the pulmonary valve, care being taken not to injure important coronary arteries.

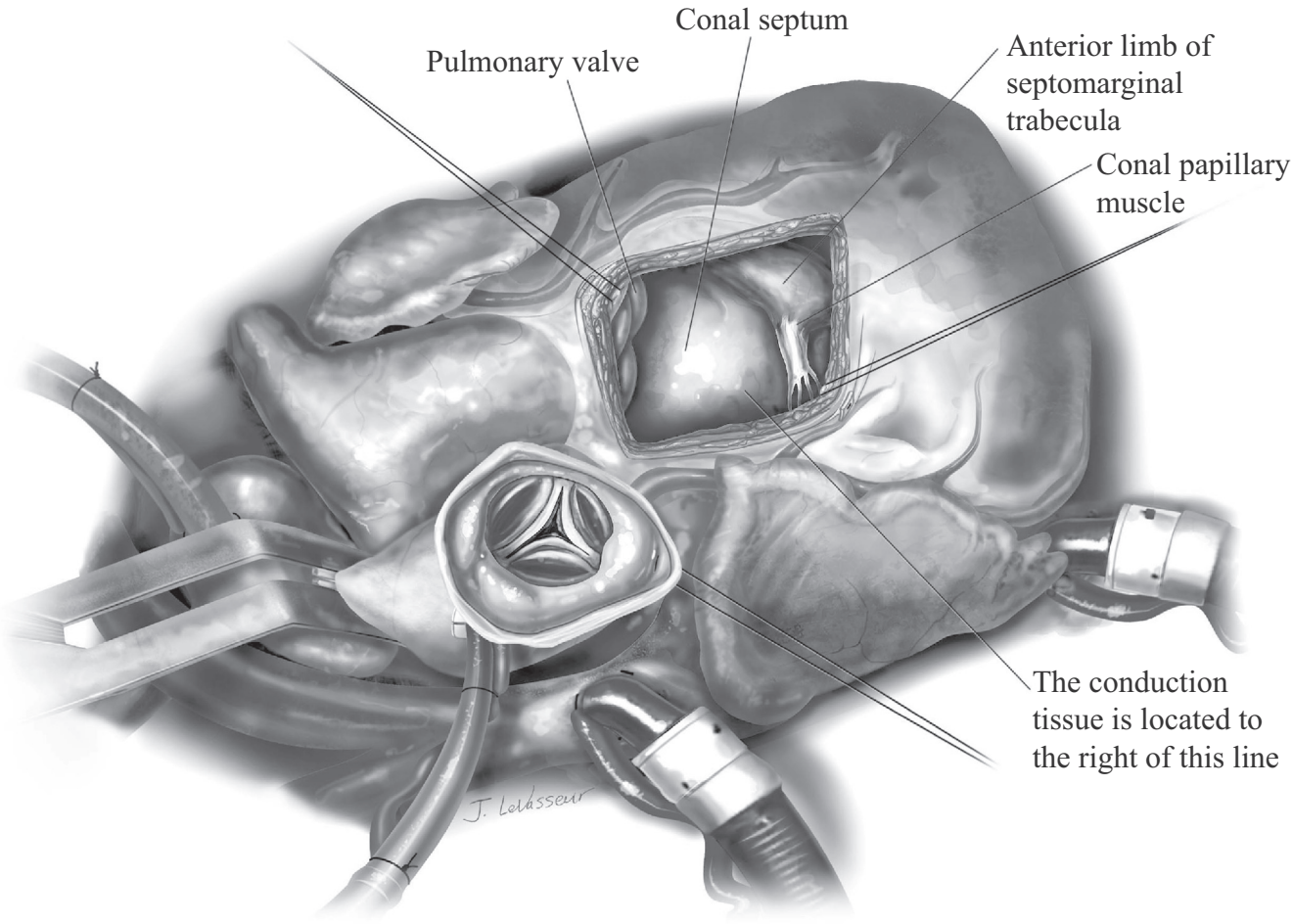

Figure 2 The right ventriculotomy is extended with direct visualization of the pulmonary valve to avoid injury to it. Once the conal septum is exposed, the region of the conduction tissue is identified, located to the right of a line coursing between the nadir of the right coronary cusp and the septal attachments of the septal leaflet of the tricuspid valve (the conal papillary muscle).

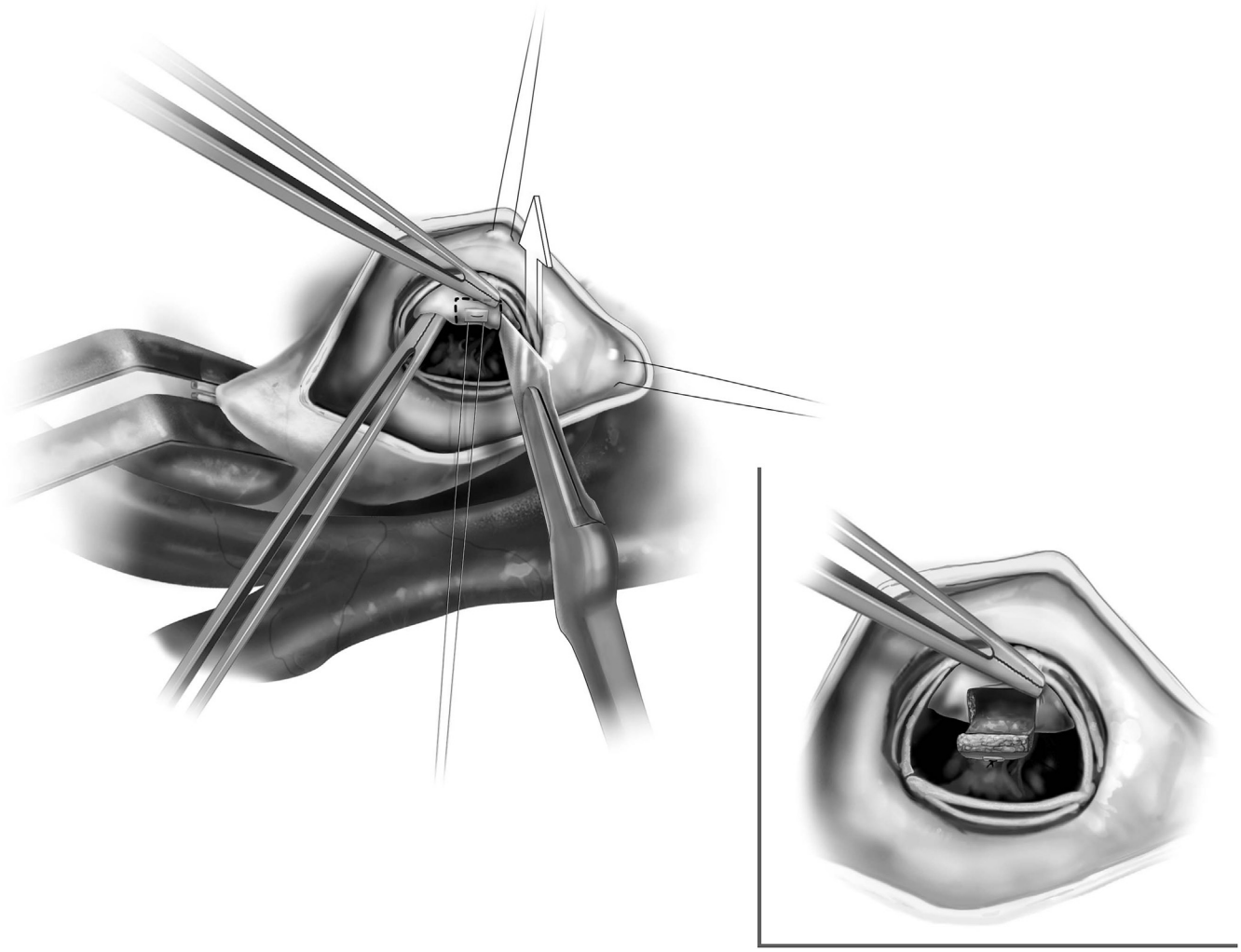

Figure 3 Working through the aortic orifice, the top of the subaortic obstruction is incised. This step is essential to achieve adequate resection of the upper part of the stenosis and to prevent damage to the aortic cusps during the subsequent surgical steps.

After gentle retraction of the aortic cusps, a traction pledgeted suture is placed on the protruding ventricular septum. Two parallel longitudinal incisions are made in the septum: the first beneath the nadir of the right coronary cusp and the second beneath the commissure between the right and the left coronary cusps. These incisions are connected superiorly with a third incision a few millimeters below the aortic valve. A deep incision is made in the ventricular septum. The traction suture is tied and left in place for later localization.

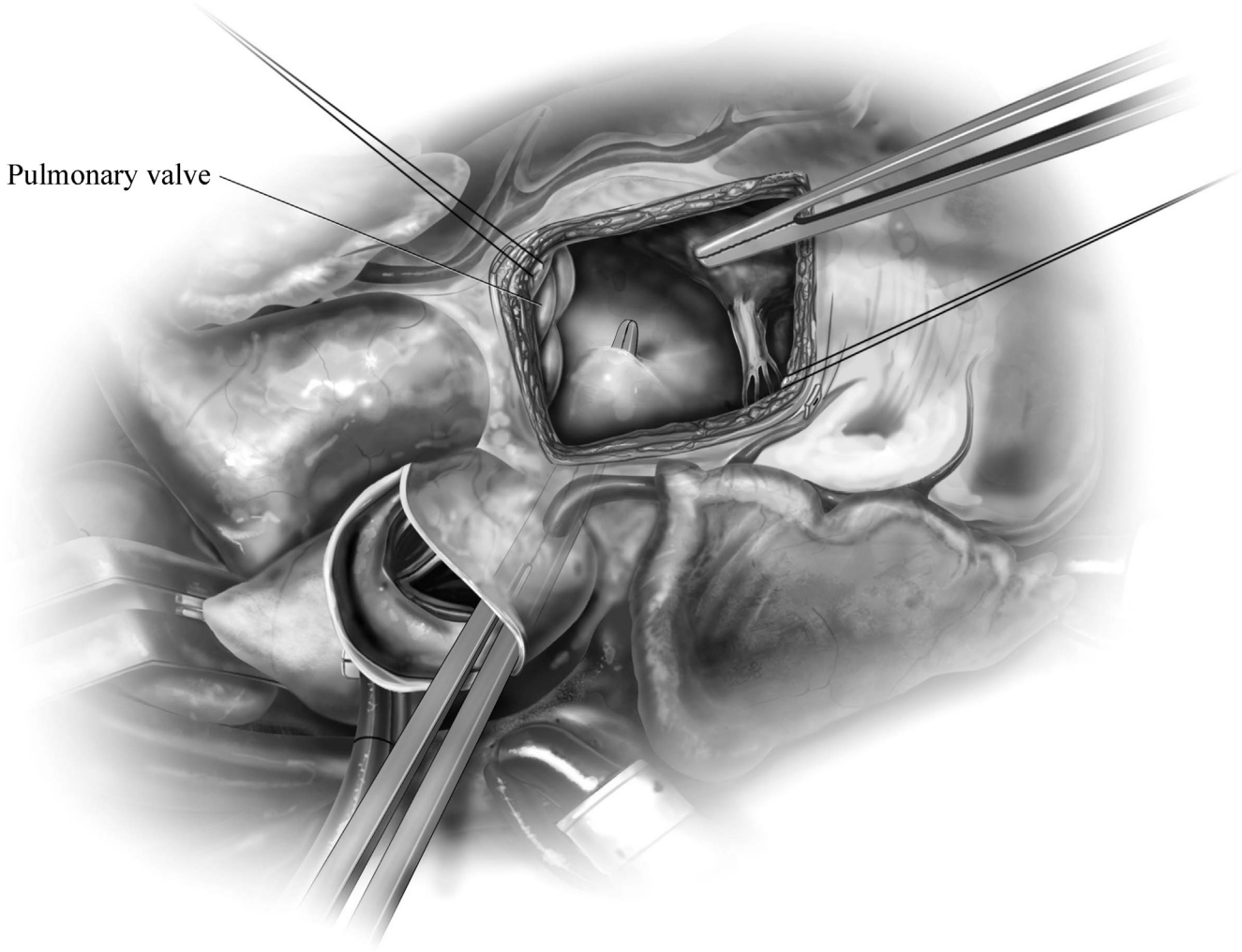

Figure 4 A right-angle instrument is introduced through the aortic orifice into the septal wedge, and the tip of the clamp is used to perforate the conal septum. Great care is taken to remain away from the conduction tissue, which has been previously localized.

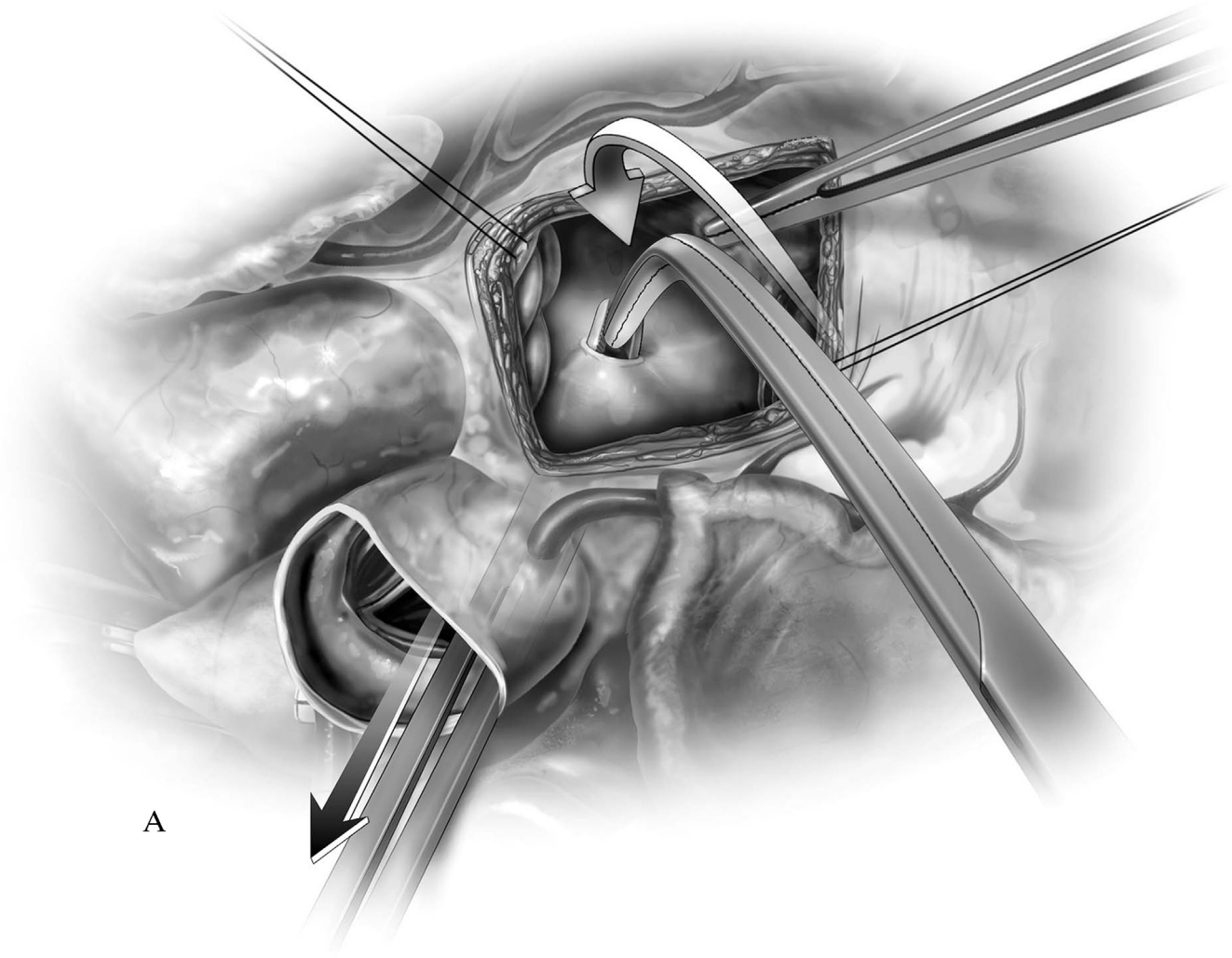

Figure 5 Another right-angle instrument is used to perform a safe and adequate incision of the conal septum.

(A) After perforation of the conal septum with the first right-angle clamp (from the left ventricle to the right ventricle), the other right-angle clamp is passed through the same hole in the other direction (from the right ventricle to the left ventricle).

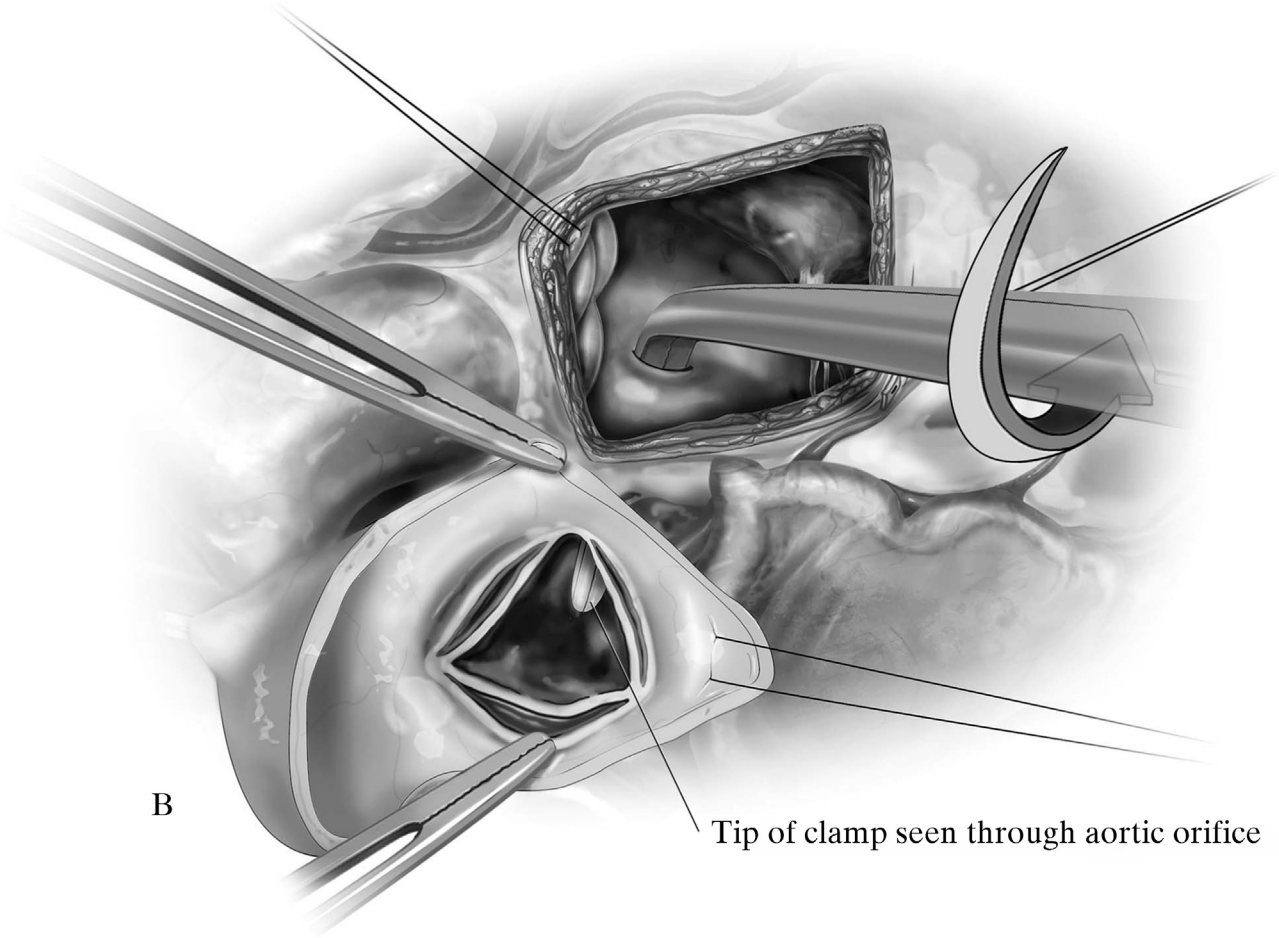

Figure 5 *(Continued)* (B) The correct position of the tip of the second right-angle clamp in the left ventricle is ascertained through the aortic orifice. The instrument is then turned 180° counterclockwise and its tip is directed toward the apex into the left ventricular cavity. The instrument is used to expose the conal septum and protect the mitral valve.

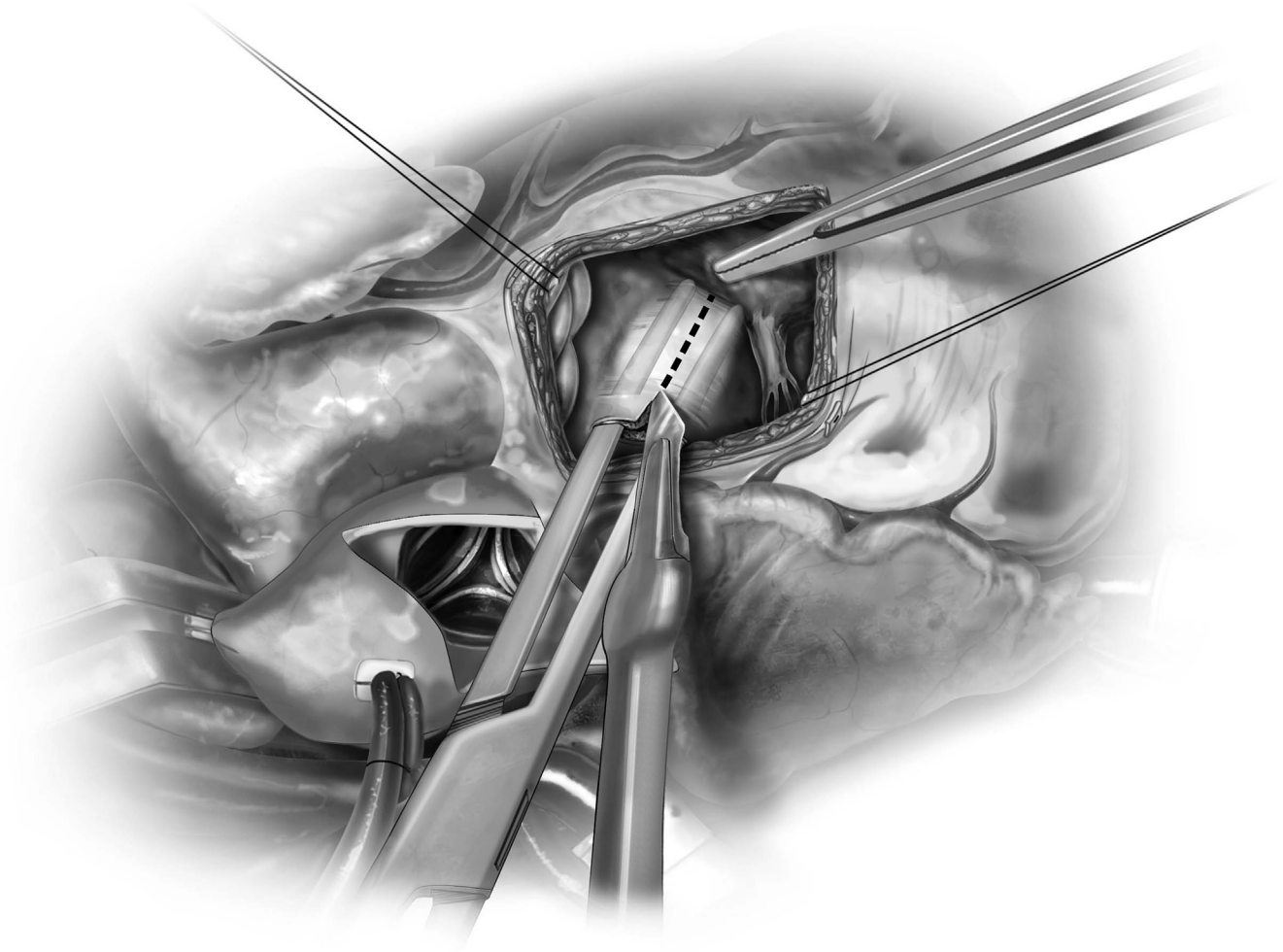

Figure 6 The right angle that has been introduced into the left ventricle is opened slightly. The conal septum in incised between the 2 jaws of the clamp.

It is essential (1) to incise the conal septum well to the left of the conduction tissue (located to the right of a line joining the nadir of the right coronary aortic cusp to the conal papillary muscle of the tricuspid valve), (2) not to extend the incision upward from the initial perforating point (to avoid damage to the aortic cusps), (3) to extend the incision downward, as far as necessary, to reach the free left ventricular cavity, and (4) to avoid dividing completely the anterior limb of the septomarginal trabecula (the biggest septal coronary artery usually courses in the lower border of this structure).

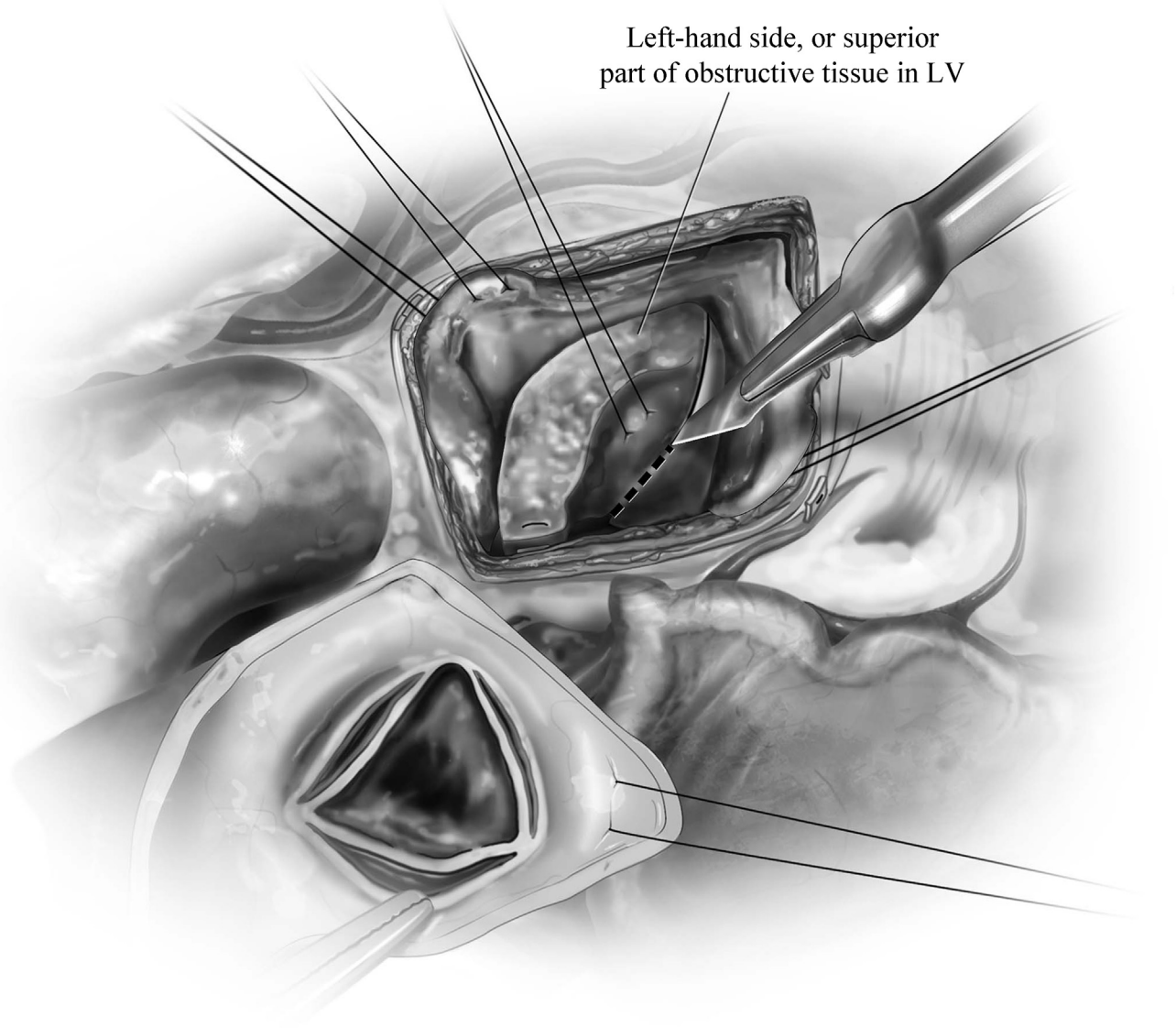

Figure 7 Two traction pledgeted sutures are placed on each rim of the septal incision on the right ventricular side and used to open the septotomy.

Resection is first performed on the left-handed (superior) side. Another traction pledgeted suture is placed on the left ventricular side of this rim. Extensive resection is carried out as much as possible, by removing the septal tissue in a single piece. The upper part of the septum (which has been incised initially through the aortic orifice and can be identified by its pledgeted suture) should be removed during this step of the resection. Even if the resection must be extensive, care should be taken to avoid damage to the free wall of the left ventricle.

Divided septal coronary arteries (easily identified during cardioplegic infusion) must be sutured to avoid the postoperative development of coronary-to-left ventricle fistulas. LV = left ventricle.

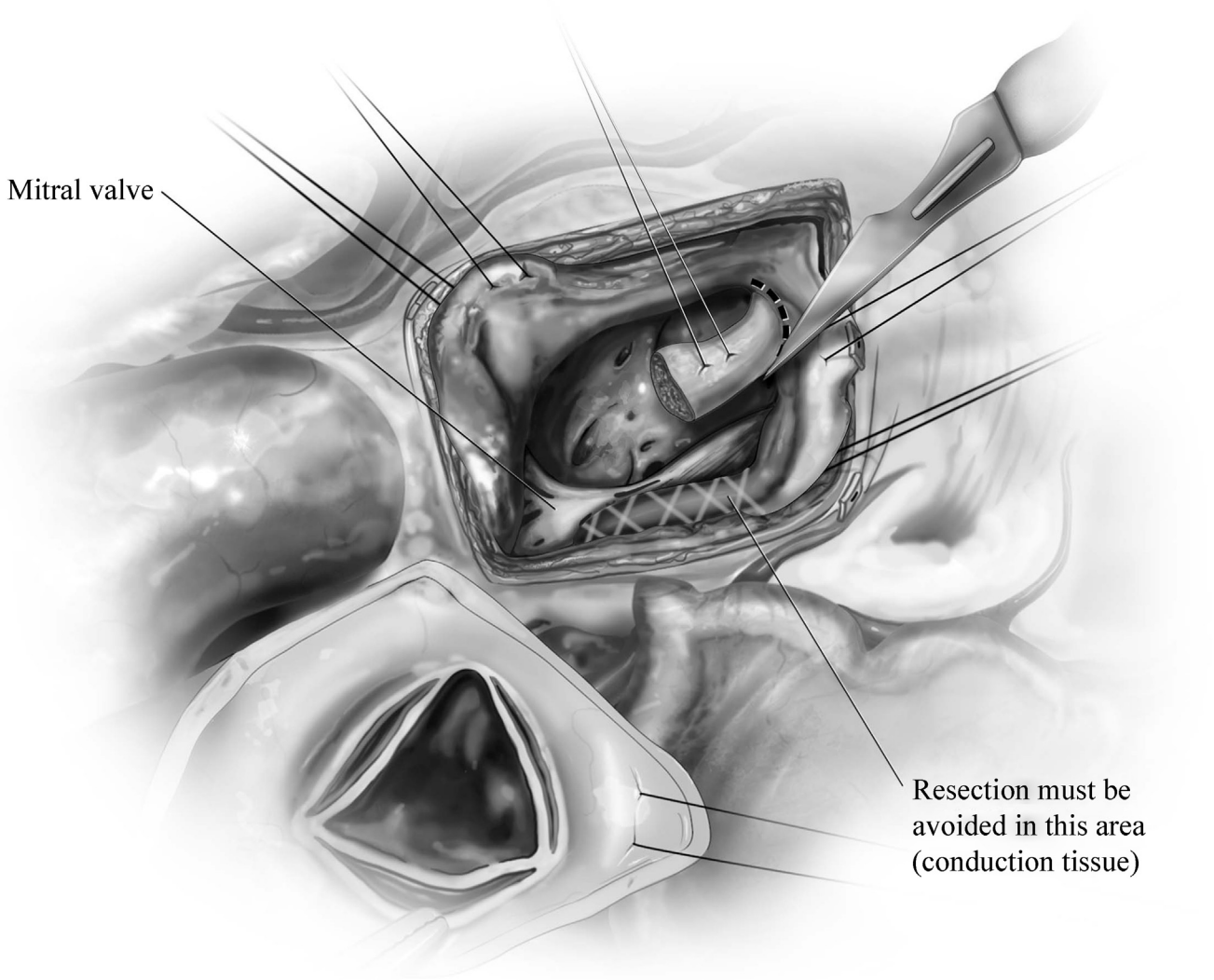

Figure 8 Septal resection is then performed on the right-handed (inferior) side. To avoid injury to the conduction tissue, it is limited to the distal part of the septotomy.

A perfect opening of the left ventricular cavity must be obtained. In most cases, additional resection must be performed; this is often the case in the distal part of the septal incision, toward the left ventricular apex. The resection must go apically well beyond the point of mitroseptal contact (usually marked by fibrous friction lesion). The adequacy and distal extension of the resection are evaluated by direct inspection and digital palpation in older children.

The mitral valve must be fully exposed and inspected. Associated anomalies of the mitral valve are addressed. Anomalous chordal attachments of the mitral leaflets to the ventricular septum or free wall are excised. An anomalous papillary muscle inserted directly onto the aortic side of the anterior mitral leaflet is divided. Care is taken to preserve all chordae attached to the free edge of the mitral leaflet and prevent the flail leaflet.

The left ventricle is irrigated to remove any particulate debris. Inspection through the aortic orifice is performed to eliminate any residual obstruction (particularly between the aortic annulus and the septal incision) and ascertain the integrity of the aortic valve.

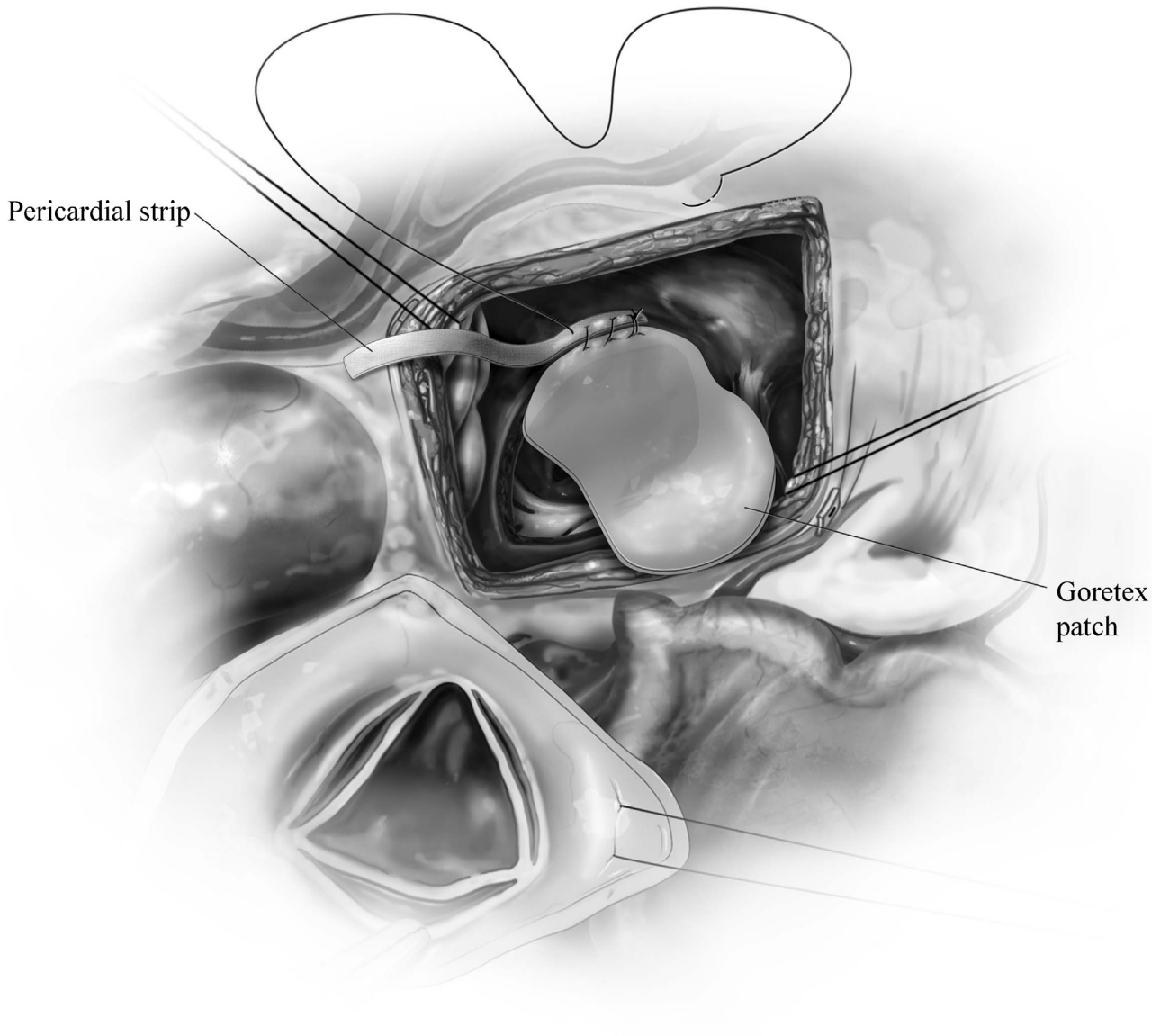

Figure 9 The septal incision is closed using a prosthetic (PTFE) patch to maintain adequate widening of the LVOT. The patch is sutured onto the right ventricular side of the septal incision, using interrupted running sutures, reinforced by pericardial strips.

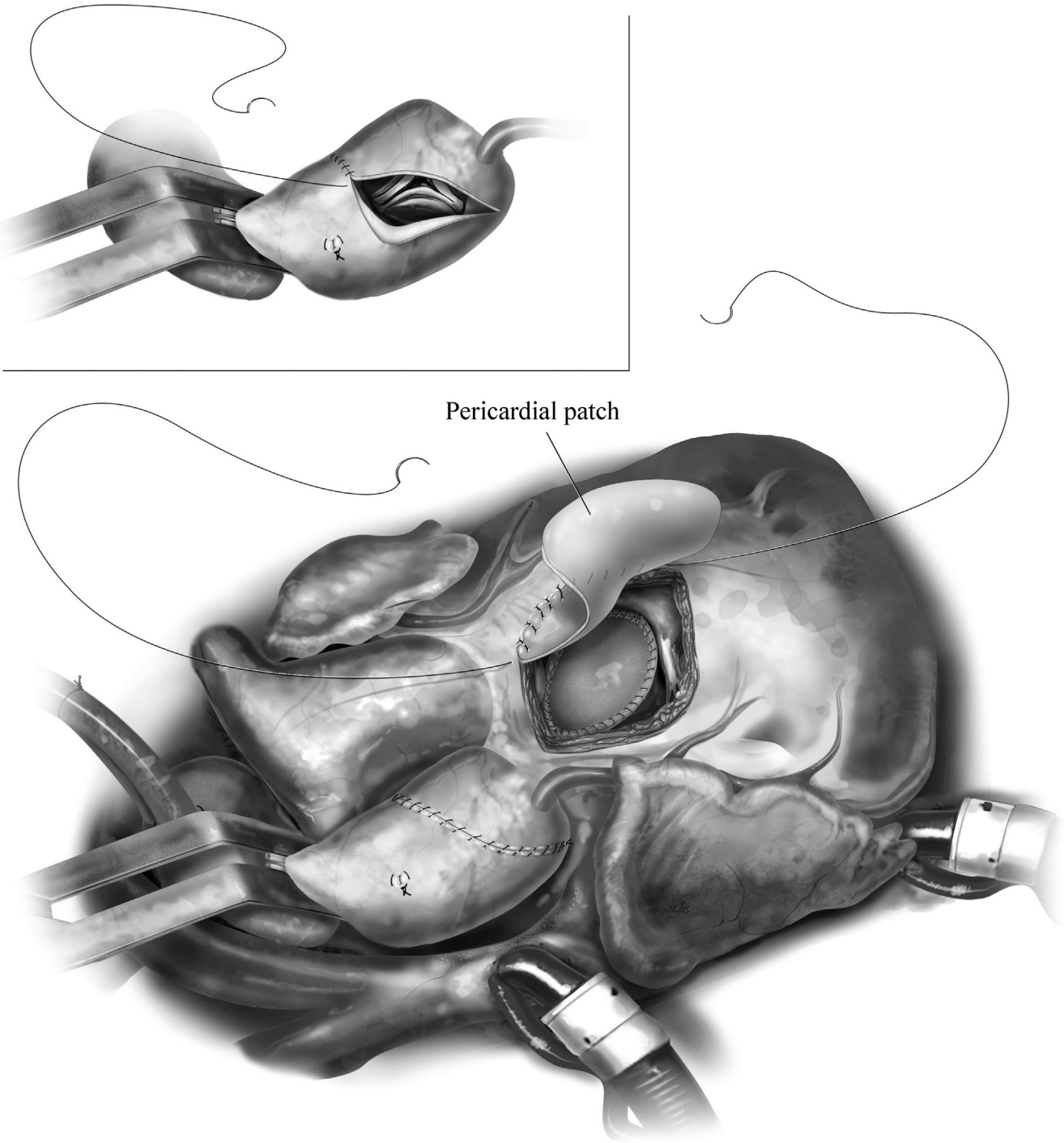

Figure 10 The aortotomy is closed after a final inspection of the LVOT. Muscular obstruction of the right ventricular outflow tract is corrected by extensive resection, if needed; this is usually the case in children with Noonan syndrome. The right ventricle is closed with a generous patch of heterologous pericardium to prevent residual subpulmonary obstruction.

Immediately after weaning from cardiopulmonary bypass, transesophageal echocardiography (TEE) is performed to assess the adequacy of the relief of the LVOT obstruction and exclude residual iatrogenic lesions (ie, ventricular septal defect, aortic insufficiency, and mitral regurgitation).

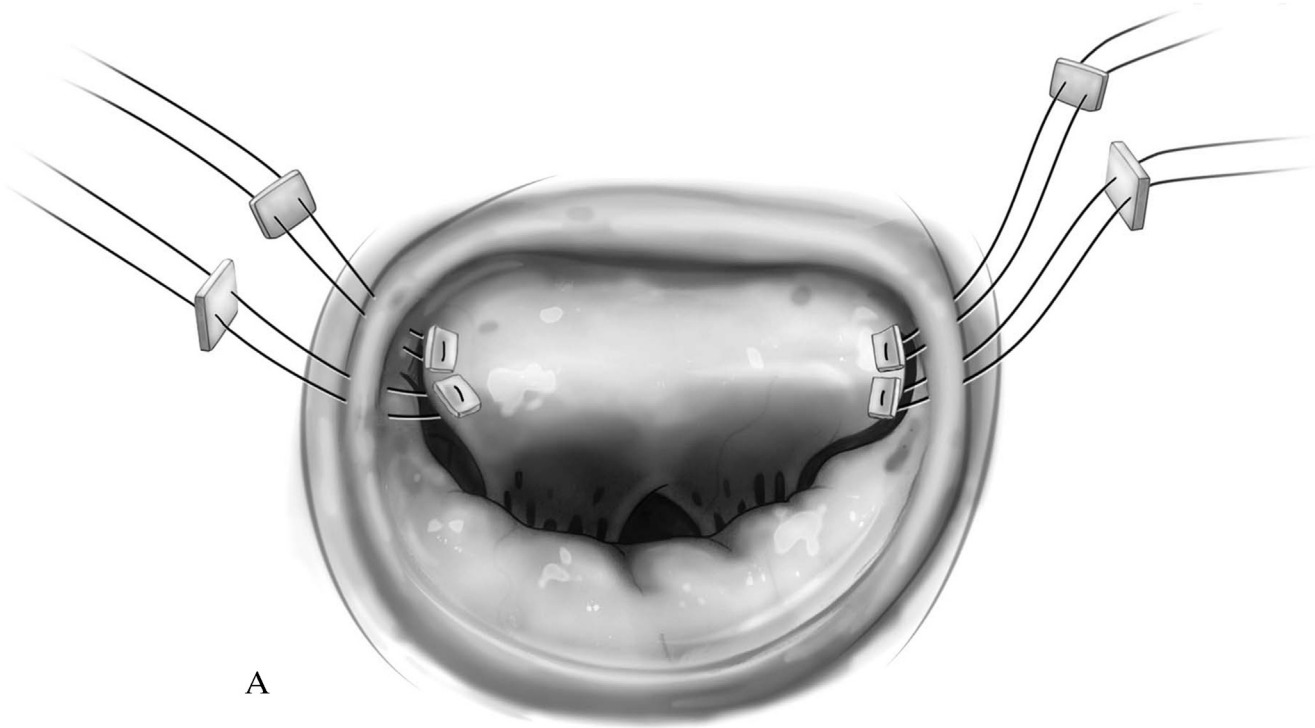

Figure 11 In most cases, associated systolic anterior motion (SAM) of the mitral valve decreases after adequate septal resection. However, there are some patients with severe SAM, usually with significant mitral regurgitation, in whom SAM may persist or worsen after septal myectomy. Associated mitral surgery may be necessary. The indication for mitral valvuloplasty may be taken before surgery or immediately after the modified Konno procedure, at the time of TEE evaluation.

In children, our procedure of choice to repair SAM is the retention plasty, as described by Delmo Walter et al.[10] A left atriotomy is performed and the mitral valve is inspected. The procedure addresses the excessive mobility of the anterior mitral leaflet by stretching the central portion of the leaflet. Mattress sutures pledgeted with autologous pericardium are used to suture the segments of the anterior mitral leaflet closest to both trigones, to the corresponding posterior annulus. The sutures are passed through the coaptation line of the anterior leaflet and the corresponding annulus of the posterior leaflet. The procedure limits the motion of the anterior mitral leaflet, thus relieving SAM and mitral regurgitation. TEE = transesophageal echocardiography.

B

Figure 11 *(Continued)* (B).

Results

Between 1990 and 2013, 51 children with HOCM underwent a valve-sparing Konno operation. The mean age at operation was 7.1 years (range: 3 months-18 years); 5 patients (10%) were younger than 1 year and 13 (25%) were younger than 2 years. A total of 12 patients (24%) had Noonan syndrome and biventricular outflow tract obstruction. Overall, 3 patients had previous surgery (septal myectomy in 2 and implantable cardiac defibrillator in 1). In association with the modified Konno procedure, 5 patients had undergone mitral valvuloplasty to correct a severe systolic anterior motion (retention Hetzer technique), and an epicardial cardiac defibrillator was implanted in 3.

There were 2 in-hospital deaths (3.9%). Both deaths occurred in infants (4 and 8 months old) with Noonan syndrome and extremely severe biventricular obstruction, and death was due to multiorgan failure (day 7 and day 45).

Postoperative complete heart block requiring permanent pacemaker occurred in 6 patients (12%); normal sinus rhythm resumed during follow-up in one of these patients. There was no complete heart block in the last 21 patients (since 2007). Heart block may be due to direct injury to the conduction tissue itself. However, in many cases, it is likely related to the division of septal arteries which participate to the vascularization of the conduction tissue. Avoiding a complete section of the anterior limb of the septomarginal trabecula should reduce this risk.

The mean follow-up was 9.3 years (range: 6 months-23 years). There was 1 late death (2.0%) (sudden death 6 years postoperatively). There was no reoperation for recurrent or residual subaortic obstruction. Overall, 5 reoperations (10%) were necessary: automated implantable cardioverter-defibrillator implantation (2), closure of residual ventricular septal defect (1), mitral valvuloplasty (1), and tricuspid valvuloplasty (1).

Summary

The valve-sparing Konno procedure provides adequate and long-lasting relief of LVOT obstruction in the pediatric population with HOCM, even in infants and small children. Mortality is low (although a word of caution should be given regarding infants with Noonan syndrome and biventricular obstruction). Iatrogenic lesions are rare, except postoperative complete heart block (the risk of which should be decreased by adequate technical details). Whether this procedure reduces late mortality and provides long-term results superior to that of transaortic myectomy remains to be determined.

Appendix. Supporting information

Supplementary data associated with this article can be found in the online version at doi:10.1053/j.optechstcvs.2014.06.001.

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